VOLUME XII August 1956 NUMBER 8

Clinical Proceedings

OF THE CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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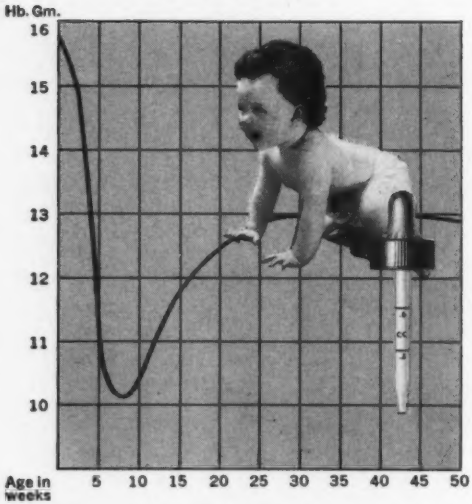

Chart adapted from Niccum, Jackson and Stearns: A.M.A. Am. J. Dis. Child. 86: 553, 1954.

Mean hemoglobin determinations of infants on a predominantly milk diet receiving Fer-In-Sol. The Fer-In-Sol dose was 5 mg. of iron (0.2 cc.) per day at 3 months, increased to 10 mg. of iron (0.4 cc.) after 6 months of age.[1]

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MEGACOLON

(Weekly Case Conference)

Reginald S. Lourie, M.D.*, W. S. Anderson, M.D.†, Frederic G. Burke, M.D.‡, I. Lattman, M.D.§, W. Pacosa, M.D.¶

A case of megacolon is presented as a focus for discussion of the several types of such a case and the psychiatric implications of this and other rectal disorders in children.

CASE REPORT

R. J., a 12 year old colored boy was admitted to Children's Hospital (third admission) March 7, 1955, for severe constipation and abdominal distension. For one week prior to this admission, constipation was more severe than usual in spite of laxatives and enemas. Onset of abdominal distension occurred two days prior to admission; this progressed to a marked degree with respiratory embarrassment on admission.

This child had been born at home, and weighed 8 pounds. He was taken to another hospital at the age of two days because of difficulty voiding; he was catheterized in the Out Patient Department and discharged.

His toilet training began at six to seven months of age and was completed at the age of one year; there was no urinary or fecal incontinence; bowel habits were normal until age five to six years when constipation started; he has since required enemas and/or laxatives two to three times a week, without which defecation would not occur for seven days or longer.

He was previously admitted to Children's Hospital, February 1951 and June 1953, with symptoms similar to those present at admission. After each admission bowel habits improved for several months but soon lapsed to the previous state. Discharge diagnoses were megacolon, fecal impaction and spina bifida occulta.

The patient is the youngest of eleven children. His school work has been "average". He is shy about going to the toilet at school where "he may be seen by other boys". The family frequently remind him of the need to have a bowel movement.

The patient's mother, father, and ten siblings are all well. The mother had trouble with constipation at one time.

On physical examination the temperature was 99°F., pulse 100 and respirations 36. He was in acute distress with shallow respirations. There was abdominal pain on each inspiration; the abdomen showed marked distension and tympany; no definite masses or organs were palpable. Rectal examination showed good sphincter tone. The rectum was filled with impacted feces. No neurological abnormality was noted. The fecal impaction was broken, manually, and this procedure was followed by several copious liquid stools and relief of symptoms.

While in the hospital the patient was noted to be conforming and interested in

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approval. No spontaneous bowel movements occurred during the hospital stay nor could satisfactory cleansing be obtained even with repeated purgatives and enemas. His appetite was good. No urinary difficulty was noted.

DISCUSSION

Dr. Anderson:

The present case is illustrative of the general problem of megacolon. Megacolon means an increased diameter of the colon with hypertrophy of both the circular and the longitudinal muscles. The hypertrophy and dilatation is due to obstruction in the distal portion of the colon and the degree is related to the duration and the completeness of the obstruction. The entire colon and maybe even the ileum may be involved. Two of the three types that are described are: those due to mechanical origin such as an anal-rectal malformation or cicatricial stenosis; and secondly, psychogenic obstipation where the anal sphincter is usually the site of the obstruction. In both of these two types, of course, the neural mechanisms are intact. In the psychogenic type the anal sphincter may or may not be spastic but a megarectum is the rule. Treatment of these cases is mainly symptomatic using such measures as mineral oil or a mild laxative to prevent solidification of the stools. Daily cleansing enemas may prevent obstruction or eventual ulceration. In the mechanically obstructed type surgical treatment to correct stenoses may be indicated. The psychogenic obstipation type will be discussed by Dr. Lourie. The third type, which is entirely different, is the so-called idiopathic megacolon or Hirschsprung's disease wherein the cause of the obstruction is not always obvious, and gross examination usually fails to show an actual mechanical obstruction. The colonic hypertrophy and the dilatation however, may be very extreme, and this type is a good deal more common than the mechanical type. Hirschsprung described this condition in 1886 and up until as recently as 1940 the disease process was thought to be in the dilated colon itself as a neuro-muscular imbalance. Then in 1948, Swenson and his co-workers showed evidence that the anatomic lesion was a congenital lack of intrinsic autonomic neurons, myenteric and submucous ganglion cells being completely absent from the lower non-dilated segment. In about 90 per cent of the cases the ganglia are absent only from the rectal segment of the colon. In 80 per cent of these cases, obstipation begins actually at birth. In the older children, such as in this patient we are showing today where the difficulty did not arise until he was 4 or 5 years old, it is very important to differentiate between the congenital megacolon and one due to psychogenic obstipation. It was in 1950, I believe, that Dr. Swenson published his operative procedure on the surgical removal of the portion of the lower bowel whose innervations are defective. Today we are emphasizing the psychogenic obstipation more than the surgical angle, and we have asked Dr. Lourie to come and discuss that type for us.

Dr. Lourie:

One of the most difficult problems to deal with in practice is the problem of megacolon. One reason is that no matter what the origin of the megacolon, emotional factors are involved. Because of the fact that the child can not maintain his own pattern of evacuation he involves the parents, since he needs outside help. Once this is established there is a distorted pattern of parent-child functioning, certainly as far as bowel evacuation goes. This distortion not infrequently leaks over into the rest of the functioning between the parent and the child. There is no question that in terms of treatment, it needs to be quite definitely decided whether one is dealing with a primary or a secondary form of megacolon. It is obvious that where there is a primary form of pathology, that needs to be dealt with first. However, even when there is a primary diagnosis of Hirschsprung's disease, we are impressed by the intermittent character of the acute episodes in which such complications as impactions occur. As we examine these situations closely, we find that even where there is a true Hirschsprung's disease, emotional disturbance often comes into the picture to precipitate an acute episode. In other words, even with primary colon pathology we often find that a pattern of combined pediatric and environmental adjustment can be worked out that will operate fairly adequately as Dr. Anderson outlines. But let something come along to disturb this balance and we see the child in an acute situation with impaction, distension, and loss of control over stools, all of which need to be relieved. On this basis we have repeated hospitalizations as a not uncommon story in these cases.

We have to deal not only with children but with parents when it comes to the treatment of megacolon. The parental aspect of this syndrome not infrequently can be traced back to the parents' own concern and attitudes about bowel functions. This is particularly true in the secondary or functional form of megacolon, described by Richmond[1] as resulting from chronic obstipation usually on the basis of long standing patterns of withholding stools. In other words, even where the etiology is psychogenic, we are dealing with essentially a psychosomatic disturbance by the time it is well established; we are dealing with a dilated colon with abnormal anatomical patterns, and of course, along with it, distorted anatomical functioning.

Once the pattern is established, no matter on what basis, when a crisis occurs we need to get the child in as normal state of balance as we can. This involves breaking up impactions, and aiding evacuation with multiple means. After we get the child in fairly good balance and able to go out of the hospital we need to provide insurance that the whole process will not start all over again. It is at this point that we have to look at the etiology.

Before the fundamental etiology is clear we have to identify the *secondary values* that have grown up around the child's difficulties whether with soiling or with constipation. For example, it becomes very easy to stay out of

school on the basis of such symptoms. It becomes very easy to keep mother away from the other siblings; to keep the parents up all night; and certainly to keep the parents' thinking focussed on the child all day long, wondering, worrying, has he had a stool or is he going to have one and what are we going to do about it if by bedtime he has not produced. After these secondary values have been identified we usually need to look back in the past for more fundamental etiological factors (which also could be continuing into the present).

In the search for the basic factors of a psychogenic nature that lead to the establishment of megacolon, we can profitably scrutinize what has happened to the normal phases of personality development. For example, in the stage of *negativism*, the child had simply to include his stool and its production in his normal pattern of expressing his negativism. This can happen with almost every child at one point or another. If at such a point, a parent's primary concern is whether the child has a stool each day (and of course this concern is reinforced in our current thinking by what is heard on the radio and what is read in the newspapers about regularity) what starts out as negativism can be perpetuated and the child can build up other values. Another point at which we have seen such disturbances start, is around 18 months when the child starts *experimenting* with its stools (just as it experiments with every other part of itself), as a food or as a painting medium. This can reach a level of severe conflict between a parent and a child. For example, mother is so shocked and so upset about this kind of experimenting that she impresses the child very strongly that this is something that must must not be; this in turn can lead to the child holding on to its stools, avoiding the total process which upset the mother to such an extent.

Another aspect that comes up in the first few years of life, again involving the parental attitudes is an *over evaluation* of *bowel function*. In other words a child finds that by whether he produces a stool every day, he can make the parents happy, or he can make them sad. If he has reason for upsetting them or retaliating for some frustration it becomes very easy to establish a pattern of withholding stools as a means of fighting. In this way a child can be a perfectly lovely child, not having to be nasty or aggressive in other ways if he has found this very effective way of upsetting parents.

Another familiar pattern involving stool disturbances arises from *rivalries* or situations that involve a change of status such as a new baby coming into the home. At such times there may be regression to soiling or withholding. If the child knows that on the basis of whether he has a stool or not, the parents will shift the focus of attention from the new baby to him, why give up a good thing if it works?

Another and sometimes more complicated kind of emotional basis for witholding the stools is *fear*. The fears we can have in mind are possibly

best understood in terms of the normal stages of development. There is a normal stage around 2½ to 3½ years when children are afraid of being hurt, especially of bodily hurt. We can expect situations in which this fear involves the bowel patterns. This may sometimes be on a basis that is extremely illogical to us as adults, but very logical to the child. For example, a child is afraid of hurting himself or losing part of his body; he will not go near lawnmowers in the presence of this fear or he will not go near vacuum cleaners or dogs, because a toe or a leg might get cut off and lost. By the same token we find that the child will suddenly find that he will not want to lose his stools. This he may think of as a part of himself that can be lost and it must not be lost. Under these circumstances we see patterns of extreme straining to hold on to stools until the child can no longer hold on. We may see suddenly developing fear of the toilet itself; he will not go anywhere near the toilet. He may have been perfectly well toilet trained but with the new fear he is not going to get into the situation that will lead to his hurting himself or losing a part of himself. This can be precipitated by a period of constipation on a dietary basis or following illness when a stool will actually cause pain. The pain and particularly the location of the pain can focus the fear of hurt on this area of his body; he then can feel in double jeopardy and may avoid both the loss of stools and the production of the pain.

Fortunately most of these disturbances are transitory. Unfortunately a few children do become so concerned that they return to being more dependent, including dependency on others for bowel care. If the handling then includes enemas, laxatives and so on, the child can find it does not have to be dependent on itself any more for the production of his stools; someone else will take care of them. It is quite helpful in these matters to know about the parents' attitudes toward bowel functioning; the ways in which they were dealt with in these matters as children and what anxieties and/or pleasures they have related to their children's toileting.

You may wonder why such frequently encountered phases of development are brought into a discussion on the psychogenic aspects of megacolon. It is in these apparently innocuous, the troublesome "usual problems" that we find roots of the more profound and persistent psychological involvements of bowel function. Since the handling of the problem in its earlier phases determines what values come to be involved in the bowel patterns, it is obvious that the doctor can be in the forefront of preventing these patterns or perpetuating them. He can be helpful in prevention by creating awareness in parents of the pitfalls, by avoiding the types of dependency, unnecessary anal stimulation and focusing of attention on this area. Or else he can be a party to prescribing or condoning the practises related to bowel functioning that form traps in which the parent and child can become entangled.

Dr. Burke:

Congenital idiopathic hypertrophy and secondary dilation of the colon associated with spasm in the anorecto-sigmoid region is most commonly seen proximal to this region. The exact neurogenic imbalance has not been conclusively demonstrated but is characterized roentgenographically by an area of contraction and spasm most commonly seen in the distal end of the colon. The failure of the distal segment to dilate and relax has been shown to be associated with or due to congenital absence of the ganglion cells in the myenteric plexus. The characteristic changes of hypertrophy, dilation and redundency are secondary to the obstruction and develop progressively

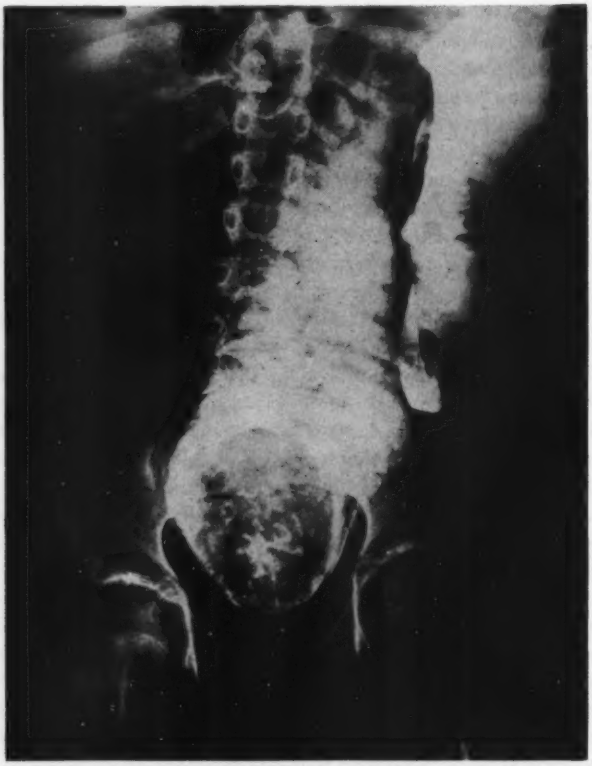

FIG. 1. Air contrast of the distal portion of the colon showing marked hypertrophy and dilation of the sigmoid with marked narrowing and spasm in the anorectal region. The spasm illustrated in this case is characteristic and diagnostic of Hirschprung's Disease.

with growth. Impaction with ischemic necrosis due to pressure changes in the bowel wall frequently lead to complications.

The demonstration of these changes is quite simply accomplished by rectal instillation of a small amount of a thin barium mixture combined with air contrast. Large amount of fluids that would be required to outline a megacolon fully should be avoided to prevent water intoxication that has resulted in collapse and death. It is wise to add a teaspoon of salt to the enema to provide hypertonicity and thus avoid this complication. Similarly, caution should be exercised in this procedure to avoid large amounts of barium that might cause a fecal impaction by concretion of the dilated colon were it to be completely filled. Probably too much barium was used in demonstrating the accompanying case but dramatic demonstration of these changes are provided in Figure 1.

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CONGENITAL ABSENCE OF THE GALL-BLADDER

Rene Jutras, M.D.*, E. Clarence Rice, M.D.†

The congenital absence of the gall-bladder is a rare anomaly diagnosed either on the occasion of a laparotomy or at autopsy. Although this condition was known to Aristotle (384–322 B. C.)[1], the first pathological report is credited to Lemery le Fils (A. D. 1701)[2]. Adding the figures published by Hagel[3], Kirschbaum[4], Knox[5], Mouzas[6], Smith[7], Stolkind[8], and Talmadge[9], one gets the following result: 62 cases in 44,320 autopsies, which gives a fair idea of the incidence of this anomaly.

The etiology of this condition is obscure. There seems to be neither sex nor racial predisposition. Cases have been reported from early infancy to old age. Bower[10], in 1928 abstracted from the literature 22 cases in infants below 1 year of age, and Latimer[11], in 1947 collected 68 cases in adults, the oldest being 84 years of age, and added three of his own. Kobacker[12], reported two cases within the same family with three other members of the family being suspected after x-ray investigation, which brings up the possibility of an hereditary factor. It is interesting to note here that this etiology was also postulated by Lemery le Fils on the ground that the hepatic disease from which his patient died was hereditary in this patient's family.

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The commonly accepted theory of the embryologic mechanism of this defect, say Hillman, Rosenthiel, Brule[13] and Malmstrom[14], is as follows: Soon after the hepatic diverticulum has emerged from the endodermal foregut in the second week of foetal life, the anlage of the cystic duct and gall-bladder starts to bud from its proximal inferior aspect. This budding may not develop at all and there will be a complete absence of the cystic duct and the gall-bladder. The study of Latimer[11], included 33 such cases. If this out-pocketing does develop as expected it will go through a normal solid embryological stage, and afterwards the lumen will be re-established spontaneously to produce a cystic duct and a gall-bladder. Failure of the latter to occur, as suggested by Gross[15], will result in a simple absence of the gall-bladder with the presence of an atretic cystic duct stump. Four such cases were listed in Bower's[10] report. By the end of the sixth week of gestation "the die is cast" and the foetus has or will never have a gall-bladder.

Mentzer[16] has done an extensive study of the comparative anatomy and found that the humans afflicted by this anomaly share it in common with 26 species of mammals for whom it is normal and among which are the horse, deer, camel and mule. The defect is optional in the giraffe and the elephant, and one must mention that all the mammals concerned are herbivorous. Dissection will also fail to demonstrate a gall-bladder in 9 species of birds, among them the pigeon, and in 17 species of fish.

The absence of the gall-bladder is often associated with other anomalies of the biliary tract, mainly the atresia of either the extrahepatic ducts or the common duct, or even the absence of the entire extrahepatic biliary tract. The latter was found in 13 of the 62 cases studied by Bower[10] with an average duration of life of 70 days. On the other hand, dilatation of the hepatic and common ducts is not infrequently found, lithiasis being the most common complication in these instances. Chronic pancreatitis and cirrhosis of the liver have been observed in several cases. Danzis[17] states that the absence of the gall-bladder is often associated with anomalies of the ears and palate, with supernumerary spleen, megacolon or imperforate anus. In our review of the literature we have found nothing to substantiate this assertion.

The symptomatology of simple agenesis of the gall-bladder (not complicated by biliary obstruction) during infancy and childhood is not established. It seems that the great majority of those who reach adult life with this anomaly will develop signs of chronic dyspepsia characterized by vague malaise following meals (Hillman, Rosenthiel and Brule[13]) or dull pain and sensation of fullness in the right hypochondrium (Latimer[11]). In their study of 60 cases published between 1900 and 1945, Dixon and Litchman[18] found that 58.8 per cent presented biliary symptoms of some sort, 50 per cent had icterus; 31 per cent had dilatation of the choledochus; 28 per cent

had lithiasis; 21 per cent had pancreatitis; and 21 per cent had cirrhosis of the liver. Of the 90 cases reviewed by Hillman, et al.,[13], 50 were diagnosed at autopsy, 36 in the operating room and 4 remained obscure as to the manner in which the diagnosis was established. In the surgical group, 25 cases had a pre-operative diagnosis of cholecystitis.

To Hillman's figures published in 1949, we would like to add two forgotten cases reported respectively by Cuynat[19], in 1838, and Godfrey[20], in 1843, and some 23 new cases collected in the available literature[6, 12, 21-30], up to 1956, thus making a total of 114. The following is believed to be the 115th and the first one reported in a patient of negro race.

CASE REPORT

V.H., a 6½ months old colored girl was admitted to Children's Hospital on December 30, 1955, with a history of severe diarrhea of 24 hours duration. She had a previous episode of diarrhea at 2½ months of age which necessitated hospital admission. On that occasion a strain of E. Coli OB:127 was isolated in cultures from the throat and from the stools. Tuberculin skin test (P.P.D. #1) and sickle cell tests were reported negative.

The family history was not contributory except that the father had been suffering his entire life from chronic digestive upsets with frequent bouts of diarrhea.

On the second admission, the infant was found to be very listless and apathetic, with a fever of 104, a pulse rate of 130 per min., respirations being shallow and rapid (40 per minute). The anterior fontanelle was depressed; the eyes were sunken; the mucosae and the skin were dry without appreciable change in the turgor. The left tympanic membrane and the pharynx were moderately congested. The remainder of the physical examination was essentially negative.

Examination of the spinal fluid was negative. The CO_2 combining power was 16 vol. per 100 ml, and the serum chloride 129 mEq/L. Attempts to correct this electrolyte imbalance and to rehydrate the patient were initiated promptly after admission. However, the infant expired 3 hours after admission.

At autopsy, the body of the 6½ months old colored infant, measuring 61 cm. in length, weighing 5.5 kilograms with an average skin panniculus of 0.7 cm. in thickness, appeared to be in a fair state of development and nutrition. The macroscopic and microscopic examinations of the brain, chest organs and abdominal viscera showed no remarkable pathologic changes. The gastrointestinal tract showed a mild congestion of the terminal ileum and the presence of a small amount of light yellow liquid stool in the colon. Heart's blood and stool cultures were negative. The only finding of interest and also entirely unexpected, was the absence of the gall-bladder and cystic duct. The hepatic and common ducts were present and drained normally into the duodenum without evidence of obstruction, dilatation or lithiasis. The inferior surface of the liver did not present any gall-bladder groove and serial section of the organ failed to reveal any intra-hepatic gall-bladder.

DISCUSSION

There is little doubt that the immediate cause of death in this infant was prolonged and progressive acidosis with rather severe dehydration. The fact that the patient had two severe bouts of diarrhea within a few months,

although far from being unusual among infants, may have had some relationship to the absence of the baby's gall-bladder.

In the patient whose gall-bladder has been removed, one of the factors for maintaining a balanced pressure relationship in the extra-hepatic biliary tree is eliminated. The sphincter of Odi, which normally relaxes in response to contraction of the gall-bladder may remain in a state of increased tonus in the absence of this stimulus for relaxation. The increased tonus of the sphincter may result in a dilatation of the common duct and in some cases the sphincter may remain in spasm, says Spellberg[21]. Not all writers agree with this statement.

It is conceivable that this anomaly may have caused an abnormal emptying of the bile into the duodenum of this patient and this might have contributed to the impairment of the digestive process. Possibly such a patient might have had a predisposition to recurrent enteritis due to the possible variation in the rate of flow of bile from the liver due to the agenesis of the gall-bladder.

It would be desirable to investigate the parents' biliary tracts, particularly the father, who gives the history of repeated gastro-intestinal disturbance since infancy, in view of the possibility of absence of gall-bladder being a family trait, as postulated by Lemery le Fils in 1701 and Kobacker in 1950.

SUMMARY

Absence of the gall-bladder in a 6½ months old baby is reported. This is believed to be the one hundred fifteenth case to be reported and the first in a negro.

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CLINICAL PATHOLOGICAL CONFERENCE

E. Clarence Rice, M.D.*, Grace H. Guin, M.D.†, Neville Connolly, M.D.‡

PROTOCOL

This 3 months old colored boy was admitted to the hospital with the chief complaints of vomiting and progressive abdominal distension of eight hours duration. Past history revealed that the infant had never seemed to thrive normally. He was the product of a full-term normal pregnancy and normal delivery. Birth weight was 2.9 kg. He was breast fed and was given supplemental feeding with an evaporated milk formula. He was seen by a physician at 6 weeks of age for a regular check-up but no comments were made to the mother as to his state of health. His mother stated that although he occasionally vomited, he had always had normal stools. He began vomiting on the day of admission but had not had diarrhea. His temperature was not elevated.

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Family history was non-contributory.

Physical examination on admission revealed a small, undernourished colored boy who appeared quite ill. He was moderately dehydrated. His rectal temperature was 98.2°, heart rate 140 per minute and respiratory rate 36 per minute. His skin was dry and scaly, had poor turgor and little subcutaneous fat. The anterior fontanelle was depressed. His abdomen was moderately distended.

Admission CBC revealed a hemoglobin of 11.4 gms., a white blood count of 11,400 with 62 per cent segmented cells and 38 per cent lymphocytes.

Initial therapy included intravenous fluids, intravenous hydrocortisone hemisuccinate (Solu-Cortef) (100 mg.) and subcutaneous coramine. Roentgenographic examination of the abdomen was suggestive of intestinal obstruction. The infant appeared desperately ill throughout his entire hospital course, vomited bile stained material several times, and expired 13 hours after admission.

DISCUSSION

Dr. Connolly:

The first thing that impressed me about this case was that the information we have about the infant's past history is so incredibly sketchy that we had best pay very little attention to it. He was breast fed but was obviously thought not to be getting enough food because he was given a supplementary formula; there is no information about supplementary vitamins. He was seen at age 6 weeks by a physician who gave the mother no clues as to the state of her infant's health; one can only assume that the child was perfectly normal at 6 weeks of age. He had never seemed to thrive normally, was regurgitating some of his feedings, was not gaining weight properly, was having episodes of crying which suggested he was having recurrent abdominal pain. I can not really make anything out of this history.

This child started to vomit 8 hours before admission and died 13 hours afterward, so something catastrophic happened in that time. I think that when you are confronted with a problem such as this you have to go through the whole gamut of diagnoses. First of all, let us consider a congenital abnormality. So far as this is concerned our attention is concentrated on the intestinal tract since this child had been vomiting and was admitted with the physical findings of abdominal distention. I am first going to consider what kind of congenital abnormality could possibly be present in the intestinal tract. We can rule out anything in the way of an intestinal atresia because the child lived 3 months, and these infants, if untreated, usually die in the neonatal period. There are a variety of other abnormalities which may cause trouble later on. In this respect I would think first of a malrotation of the gut or malfixation of the mesentery. These conditions will usually

present with colicky abdominal pain indicative of an intermittent intestinal obstruction which may be high or low according to where the lesion is. The most common type of a malrotation with malfixation of the gut gives symptoms as a result of a congenital band lying across the duodenum and attached to a large intestine which is not rotated into the iliac fossa. The obstruction, if it occurs, is distal to the pylorus, and if of long standing, the pylorus becomes incompetent. Since the common bile duct enters the duodenum proximal to the obstruction, any vomitus is usually bile stained. Intermittent duodenal obstruction from such a congenital band is a definite possibility in this case. At anytime, the loop of small intestine caught in this band may rotate around, cut off its venous drainage even if it does not affect its arterial blood supply, and produce acute gangrenous obstruction of the involved segment of intestine. This will produce a blind loop intestinal obstruction which is an invariably lethal condition.

Another condition to be considered is that resulting from a remnant of the vitelline duct attached from a Meckel's diverticulum to the umbilicus. This is really just a band inside the abdomen around which the intestines may get twisted. This may occur at any period during a person's life or may never occur. There are two other congenital abnormalities which can produce rather bizarre pictures. One is a congenital absence of the myenteric plexus giving rise to Hirschsprung's disease. You may think it silly to discuss Hirschsprung's disease in reference to this particular case but we know so little about the history that I think we ought to consider it. The symptoms of Hirschsprung's disease are classically very easy to diagnose. The child is extremely constipated, and gets more and more distended. The terminal picture can be not unlike this if it occurs in infants. A rectal examination will usually rule it out because if the rectum is full of feces, it is not a typical Hirschsprung's disease. It may be somewhat atypical, but normally in this condition the dilatation of the large bowel occurs in the normal gut, proximal to the abnormal segment. The abnormal segment is in the rectosigmoid region classically and it may extend much further or it may be quite short. This abnormal segment is not contracted; it just is not dilated because it is not functioning. There are no peristaltic waves traversing this area. In essence, the intestine is trying very hard to drive everything through a noncooperating piece of tubing. It is not entirely inert; it is just contracting all over the place at the wrong time in an uncoordinated manner. The picture, then, is one of distension, and children, particularly small infants, can get extremely ill quite quickly.

The other congenital abnormality which I wish to discuss is duplication of the intestine. This abnormality again does not always manifest itself at birth. There is no reason why it should, because the fluid in the duplicated loop increases in volume and gradually causes an obstruction. It can also

lead to an intussusception. It has been reported that a small duplication of the ileocecal valve will intussuscept into the cecum and give rise to a mechanical obstruction.

However, in dealing with these congenital abnormalities, and knowing as little as one does about this child, without seeing the x-rays at any rate, you have got to think that this congenital abnormality may not be in the gut at all. A child with chronic urinary obstruction may be admitted to the hospital looking like this, and can die very quickly.

Having dealt very rapidly with congenital abnormalities, the second group of things I would like to discuss is trauma. I do not mean that the mother dropped the baby on the floor or anything like that, but trauma to my way of thinking, means not only extrinsic but intrinsic, trauma. After all, this baby, like every other baby, was born, and this of course is one of the most traumatic experiences in the entire life of an individual. Pressure such as will never be exerted again is exerted on these infants and suddenly it is let go. If the infant is born head first, the head is suddenly relieved of this tremendous pressure, and has to make a very considerable hydrodynamic adjustment. It does not seem very surprising to me that such things as subdural hematomas occur. A diagnosis of subdural hematoma would account for quite a number of things in the protocol. The infant did not thrive very well, and yet no definite abnormality was found. This is frequently the history in infants with subdural hematomas. This infant started vomiting, we are told only eight hours prior to admission, but of course it may have been in pretty poor shape before this.

Intrinsic trauma also includes the mechanical obstructions most of which have been listed under congenital abnormalities. The infant may have had a congenital band with volvulus or an intussusception. These will all result in an intestinal obstruction on a mechanical basis.

Intussusception itself can be due to a variety of things. Far and away the most common is what we choose to call idiopathic. Of all the cases of intussusception, in less than ten per cent will a demonstrable cause be found. In that small group a Meckel's diverticulum is probably the most important. Another is reduplication of the bowel which I have already mentioned. Tumor of the bowel, usually a polyp, may be a cause. A purely mechanical cause is hemorrhage into the wall of the intestine as may be seen with a blow to the abdomen, a blood dyscrasia or anaphylactoid purpura. I have operated on one infant with intussusception resulting from this latter condition.

Not all intussusceptions progress into the large bowel. Some of these odd ones, caused by such things as a hematoma or tumor may be an ileo-ileal or a jejuno-ileal intussusception, and in those cases a barium enema will be no help at all. Not only is it not therapeutic, it is not even diagnostic because the barium will fill the large bowel and in some instances may even start

to fill the small bowel. The x-ray department will then report no intussusception. What they mean is that there is no intussusception into the large bowel, but there may still be an ileo-ileal one.

Could this child have had an infection? There again, if we do not pay too much attention to the fact that the child was not thriving beforehand, I think that this child could very easily have had an infection. He could have had a primary peritonitis. Primary peritonitis, some people think, is commoner in girls than it is in boys, and results from ascending infection of the genital tract, but the evidence for that is non-existent. It is just about as common in boys as it is in girls. It has recently become considerably less common. The commonest organism is the streptococcus, and with the diminution of acute streptococcal upper respiratory infections, it is becoming a relatively rare disease. In any event, primary peritonitis may present as a very fulminating disease in a child this age, a very sick child who is very rapidly going downhill. The white count, you may argue, is against it, but not all infants react typically with a rapid rise of total white count, with an overwhelming infection, and it is dangerous to exclude a diagnosis merely because the total white cell count is not abnormally high.

Appendicitis is a rare entity in this age group but it occurs, and sufficiently often in this age group to keep a surgeon on his toes. If it does occur it is an extremely lethal disease and may have a very fulminating course.

Another type of infection which may give a very bizarre clinical picture is pneumonia. This disease very frequently presents with abdominal pain and other gastro-intestinal symptoms. The differential diagnosis between pneumonia and appendicitis may be very difficult. In these days of modern anesthesia and antibiotics, if presented with a child who has clinical and even roentgenographic evidence of pneumonia but who has abdominal complaints and some abdominal muscle guarding it is wiser, after a suitable period of observation, to remove the appendix rather than say "This child has pneumonia and so can not possibly have appendicitis". Many surgeons, I am sure, have duplicated my experience of removing a gangrenous appendix in a patient with definite x-ray evidence of a lower lobe pneumonia. Of course, I am not saying that every child with abdominal pain and some muscle guarding should have his appendix removed. However, it does require a lot of careful thought and examination before a definite decision can be made.

I must mention congenital hypertrophic pyloric stenosis. This child did not thrive very well; we have a poor history, and we do not know how much and how often it vomited before it was admitted. It was certainly thoroughly dehydrated. As for distention, it depends on who writes the history. Some physicians will call a child distended if there is only upper abdominal distension even though the lower abdomen is flat and soft. A child with

pyloric stenosis can look quite distended, particularly in the upper abdomen, and if sufficient dehydration is present, death can certainly occur. Of course this child vomited bile. If we can depend on that portion of the history, the obstruction must be beyond the pylorus. It may sometimes be very difficult to differentiate between the tumor of pyloric stenosis and that resulting from dilatation of the first part of the duodenum arising from obstruction due to a duodenal band. It, of course, does not matter too much since both conditions are best treated surgically.

Hernias must be considered. The inguinal hernia is a congenital abnormality. The internal hernias that occur are almost always associated with some type of malfixation and are another cause for "blind loop" intestinal obstruction. Most that I have seen have been associated with malfixation of the mesentery in some way or other. I think the classification of malfixations is a nightmare because there are so many different abnormalities that can occur. The common ones everybody knows about but there are some very, very bizarre conditions that are occasionally seen.

If I might summarize, I think that this baby died of a high intestinal obstruction, and I do not know what caused it, but I think that a malfixation of the mesentery is the most likely.

Dr. Guin:

The correct diagnosis is intestinal obstruction but the obstruction was a little lower than you placed it. The cecum was found to be 10 centimeters in length, and was folded on the proximal portion of the ascending colon. The loops were entwined and dilated, and their serosal surfaces were purplish, giving pretty much the appearance of gangrene which was proved on microscopic examination. The child died of a volvulus with acute peritonitis. The sections from the peritoneum and involved intestine showed acute inflammation. It was thought that perhaps the mesentery was a little bit longer than normal, thus allowing for this folding over and twisting. The appendix was normal. The adrenal glands were hypertrophied. I do not believe we have seen that in our other cases of acute intestinal obstruction with peritonitis. This child did not have any ACTH; I can not explain the hypertrophy unless it was a secondary response to the child's being in shock. There was only a small amount of pulmonary edema in the lungs and no bronchopneumonia. We cultured proteus vulgaris from the peritoneal fluid.

Final Pathological Diagnoses:

1. Generalized peritonitis
2. Intestinal obstruction and necrosis secondary to volvulus of the cecum and ascending colon.
3. Dolichocecum
4. Hypertrophy of the adrenals

Dr. Connolly:

This type of obstruction in adults is usually classified as volvulus of the large bowel, but the symptoms are those of small bowel obstruction. In adults and children with this condition, it is not necessary to divide the lateral peritoneum to lift up the cecum. The cecum lies free and is not attached down. It is covered on the posterior surface with peritoneum so that there appears to be another leaf of mesentery underneath it. This condition must exist before a volvulus of the right side of the colon can occur. I do not believe this lack of fixation is such an uncommon condition in children, and I wonder how much of this fixation occurs later on in life, because normally in an infant the cecum and ascending colon are remarkably mobile, much more so than in the adult.

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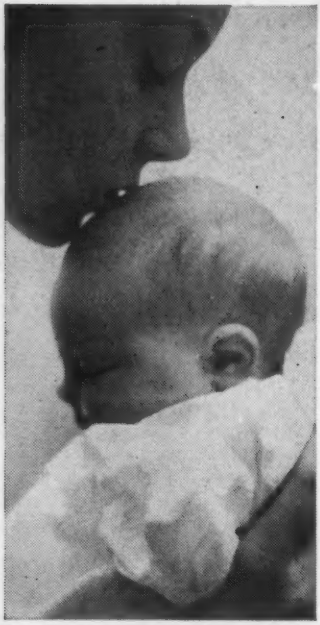

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